Congenital intrathoracic neuroblastoma presenting as persistent pulmonary hypertension in a newborn

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Clinical imaging

A female infant, weighing 2740 g, was vaginally born at 42 weeks of gestation. The Apgar scores were 7 and 8 at 1 and 5 minutes, respectively. After birth, the infant immediately developed respiratory distress, and required conventional ventilation, with an initial peak positive pressure of 15 cmH_2O, positive end-expiratory pressure of 4 cmH_2O, frequency of 40/min, and supplemental oxygen of 100%. A chest radiograph taken within the first hour following birth revealed retained fetal lung fluid with homogenous round opacity at the right upper lung (Fig. A). At 3 hours of life, she developed severe hypoxemia with 75% of pre-ductal saturation (SpO_2) and 64% of post-ductal SpO_2, leading to a clinical diagnosis of persistent pulmonary hypertension of the newborn (PPHN). Her respiration support was substantially changed to high frequency oscillatory ventilation with a mean airway pressure of 12 cmH_2O, delta-pressure of 40 cmH_2O, frequency of 10 Hz, and supplemental oxygen of 100%. The results of an arterial blood gas taken at 6 hours of life were $PaCO_2$ 79 mmHg, PaO_2 54 mmHg, and pH 7.12. Serial chest X-rays at 24 hours of life revealed disappeared fetal lung fluid and the round opacity became more obvious (Fig. B). At 3 days of life, she was weaned from supplemental oxygen, inotropic therapy, and sedative drugs, and respiratory support was switched to conventional ventilation at her 4 days of life. Computed tomography demonstrated a posterior mediastinal mass with faint internal calcifications and tracheal displacement (Fig. C). Ventilator dependency suspected to be secondary to an airway obstruction led to the attending physician performing a thoracotomy on the 12th day, which revealed a tumor which was later removed. Histology of the tumor showed small round cells in a pseudorosette formation, compatible with a poorly differentiated neuroblastoma (Fig. D). Unfortunately, the patient died of septicemia 2 months later.

Congenital neuroblastoma is the most common malignant solid tumor in newborns, accounting for 54% of all neonatal malignancies.[1] However, intrathoracic lesion is very rare in this age group. All reported neonatal intrathoracic neuroblastomas have presented with respiratory symptoms and required respiratory

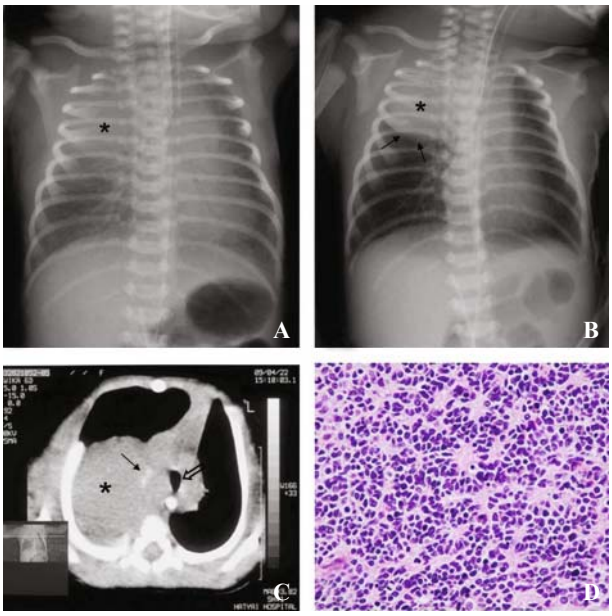

Fig. A: An initial chest radiograph showing bilateral diffuse ground-glass opacity related to retained fetal lung fluid and faint homogeneous opacity at the right upper lung (*). **B:** A serial chest radiograph revealed disappearance of fetal lung fluid. The opacity at the right upper lung became mass-like lesion (*) with bulging inferior border and tracheal deviation to the left side (arrows). **C:** Plain computed tomography image demonstrated a well defined posterior mediastinal mass (*) with internal calcifications (arrow) and tracheal displacement from mass effect (open arrow). **D:** Histopathology of the tumor showing a poorly differentiated neuroblastoma. The tumor was composed of small round cells with pseudorosette formation.

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support.[2,3] A literature review revealed no reports of association between this condition and PPHN. The pathophysiology that could explain PPHN in thoracic neuroblastoma is unclear. Underdevelopment of the pulmonary vasculature during the early gestational period, resulting in a reduction in number and size of alveoli, might be the main cause of PPHN.[4] In addition, our case suggested that follow up imaging is helpful to detect congenital intrathoracic lesion which can be missed in the initial radiographic examination due to retained alveolar fluid.

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